Atlanto - Occipital Fusion – Report Of Two Cases

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Abstract. — Skeletal abnormalities at the craniocervical junction may result in sudden unexpected death. We report two skulls showing occipitalization of the atlas. Of the two skulls, one skull showed defect in ossification called “Spina-Bifida-Posterior”. The fortuitous discovery of this skull with defects in ossification especially of the atlas prompted us to review the literature particularly on this developmental anomaly. Posterior-Spina-Bifida involves 3% of all the spines examined. Anterior and posterior foramen magnum dimensions were remarkably narrowed.

Key words : — Atlas, Foramen Magnum, Anterior Arch, Posterior Arch, Occipital Bone.

Introduction :

Craniovertebral abnormalities have been recorded for many years in morphological and clinicoradiological studies.

Occipitalization of the atlas, assimilation of atlas or atlantooccipital fusion is one of the common skeletal abnormalities of the upper cervical spine.

Hypoplasia of the basiocciput can be found in manifestations of the occipital vertebra, in atlas assimilation and in atlantooccipital fusion. These patients with craniovertebral joint anomalies exhibit the first neurological signs and symptoms usually no sooner than the second decade (Kruyff ; 1965).

Anomalies in the region of the foramen magnum were of the interest mainly to anatomists. (McRae & Bernon, 1953). These began to be described in pathological literature only in the later half of the 19th century.

Review of Literature :

Atlantooccipital fusion or occipitalization of atlas or assimilation of atlas has been extensively dealt with in the literature by craniologists. There are previous reports of both ossification defects and atlanto occipital fusion. Our study is compared with previous workers. These earlier reports are summarized below.

Table-1 Symptomatology of Atlanto-Occipital fusion.

<table>
<thead>
<tr>
<th>Sr. No.</th>
<th>Author</th>
<th>Year</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Kussmaul &amp; Tenner</td>
<td>1859</td>
<td>Convulsions in all 3 reported cases</td>
</tr>
<tr>
<td>2.</td>
<td>Englander</td>
<td>1934</td>
<td>Cervical pain</td>
</tr>
<tr>
<td>3.</td>
<td>Hadlay</td>
<td>1948</td>
<td>Neurological Symptoms.</td>
</tr>
<tr>
<td>4.</td>
<td>Keller</td>
<td>1961</td>
<td>Seizure disorder</td>
</tr>
<tr>
<td>5.</td>
<td>Lopez Zanon et al</td>
<td>1964</td>
<td>Cervical Pain in all 3 reported cases</td>
</tr>
<tr>
<td>6.</td>
<td>Budin &amp; Sondheimer</td>
<td>1966</td>
<td>with tonic clonic convulsions in two out of them</td>
</tr>
<tr>
<td>7.</td>
<td>Albert &amp; Castrischer</td>
<td>1968</td>
<td>Cervical pain following hyperextension and Onset of cervical pain while putting sweater</td>
</tr>
<tr>
<td>9.</td>
<td>Vakili et al</td>
<td>1985</td>
<td>Pain in the neck in some of 25 reported cases; others being either asymptomatic or having insufficient clinical data</td>
</tr>
</tbody>
</table>

Case Report :

In our study related to foramen magnum we noticed two skulls each showing occipitalization of atlas. These were labelled I and II.

Skull I : Showed atlanto occipital fusion with associated spina bifida posterior of the atlas. This skull showed complete fusion of only one of the transverse processes with the occipital bone. The
anterior arch of this atlas was incompletely fused with occipital bone (fig. 1).

**Skull II** : In this skull the anterior arch of atlas was fused with the occipital bone. This skull also showed fusion of only one of the transverse processes of atlas to the occipital bone. One half of the posterior arch was fused with the occipital bone and the other half of the posterior arch was probably broken (fig. 2).

**Embryology** :

In some lower vertebrates (e.g.: Reptiles) the cranial half of the first cervical sclerotome remains as a separate bone 'the proatlas' between the occiput and C1 vertebra. In man it becomes assimilated into the occipital condyles and also forms the tip of the dens. The caudal half of first cervical sclerotome gives rise to the lateral masses and the anterior and posterior arches of C1 vertebra. The body of C1 vertebra disappears early giving rise to all but the tip of the dens. The body, posterior arch and transverse process of C2 vertebra is derived from second cervical sclerotome. If the normal segmentation fails to occur, atlantooccipital fusion results. (Shipro & Robinson, 1976). The single anterior ossification of the first cervical vertebra is well described in the classic study of Macalister; 1893. According to him, “Two ossification centres appear, one just posterior to each of the lateral mass and transverse process”. The two ossification centres meet in the middle, posteriorly usually by late in the fourth year of life. At about six months of age ossification begins in the anterior part of atlas generally by two closely approximated ossification centres, which usually unite within a few months of their appearance. Occasionally there may be single anterior ossification centre. Ossification proceeds posterolaterally along both anterior and fusion to the lateral masses takes place in the fifth year of life (Macalister, 1893).

A midline defect can result from the failure of fusion of the ossification centres either anteriorly or posteriorly.

**Discussion** :

Neurological symptoms associated with occipitalization of the atlas are attributed to the ligamentous laxity of the transverse ligament about the odontoid process caused by repeated flexion and extension of the neck leading to the compression of the spinal cord or actual indentation of the medulla oblongata.

With aging, the central nervous system may be less tolerant to repeated blows from the odontoid. As per McRae’s & Barnon (1953) study in 25 patients, they noticed 2 fusions of C2 and C3 vertebrae. The patient will be probably asymptomatic if the odontoid process is located below the level of foramen magnum. This relationship is best assessed through the use of McRaes & Barnon (1953) line which is drawn across the foramen magnum. Normally the odontoid process should not project above this line.

According to Greenberg (1968) spinal cord compression always occurs when the sagittal spinal canal diameter behind the odontoid process is less than or equal to 14 mm. Cord compression is possible when the sagittal canal diameter is between 15 and 17 mm. And almost never occurs at a distance of 18 mm or more.

According to McRae & Barnon (1953), patients with occipitalization of the atlas may have the following physical features: low hairline, torticollis, restricted neck movements and / or abnormal short neck. In neurological examination of the atlanto occipital fusion patient may reveal the following clinical findings: headache, neck pain, numbness and pain in the limbs, weakness, abnormal head posture, posteriorly located dull aching headache. Cranial nerve findings associated with occipitalization of the atlas include tinnitus, visual disturbances and lower cranial nerve palsies leading to dysphagia and dysarthria. The neurological symptoms and signs of atlanto-occipital fusion can not be distinguished from those of the Arnold Chiari malformation as the pathophysiology of both is essentially the same.

Fusion between atlas and occiput occurs anteriorly between the arch and rim of the foramen with some segment of the posterior arch of C1 present in some instances. This fragment can frequently constrict the spinal canal causing intermittent symptoms depending on the position of the head. (Bailay et al, 1983).
Conclusion:

Atlanto occipital fusion reduces the foramen magnum dimension leading to neurological complications due to compression of spinal cord.

References:

Fig-1
Photograph of skull 1 showing atlanto-occipital fusion with spina bifida posterior of atlas.

Fig-2
Photograph of skull-2 showing atlanto-occipital fusion. Posterior arch of atlas is probably broken.